



Human Acanthocephaliasis: a Thorn in the Side of Parasite Diagnostics

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ABSTRACT Acanthocephala is a phylum of parasitic pseudocoelomates that infect a wide range of vertebrate and invertebrate hosts and can cause zoonotic infections in humans. The zoologic literature is quite rich and diverse; however, the human-centric literature is sparse, with sporadic reports over the past 70 years. Causal agents of acanthocephaliasis in humans are reviewed as well as their biology and life cycle. This review provides the first consolidated and summarized report of human cases of acanthocephaliasis based on English language publications, including epidemiology, clinical presentation, treatment, and diagnosis and identification.

KEYWORDS acanthocephalans, diagnosis, parasitology, epidemiology, helminths

The acanthocephalans, or thorny-headed worms, are a diverse group of parasitic worms with an estimated 1,100 species described within the phylum Acanthocephala. Because they are highly modified parasites, the relationships of acanthocephalans to other animals remains problematic, but molecular systematics suggests that they are related to the rotifers or possibly nested within Rotifera proper (1). These parasites can be found in reptiles, amphibians, birds, and mammals. The organism sizes and gross morphologies are quite different across the various members of the phylum, with some as small as 1 mm and others reaching more than 60 cm. These worms are characterized broadly by an elongated tube-like body with an anterior end containing an eversible, hooked proboscis which resides within a proboscis receptacle until contact with host tissue is made. The acanthocephalans, like cestodes, lack a mouth, intestine, circulatory system, and excretory system. Akin to nematodes, Acanthocephala are dioecious and sexually dimorphic (2).

Human cases of acanthocephaliasis are seemingly rare in medical literature; however, there have been a growing number of cases reported in the last 10 years (3–13). Previous reports have also described additional cases which were not published in full detail, which suggests that these parasites may be more frequently encountered than the medical community may appreciate (10, 14) (B. A. Mathison, unpublished data). Many popular medical microbiology textbook resources (conventionally used by diagnostic laboratories) do not include content related to the acanthocephalans. This omission from conventional text likely leads to their limited or inaccurate reporting, with many labs likely considering these worms nonparasites or identifying them inaccurately as *Ascaris lumbricoides* (7, 10).

This work represents the first consolidated review of the English language literature for the acanthocephalans, specifically focused on those species associated with human infections. Table 1 provides a summary of cases reports of human acanthocephaliasis published in English language literature.

CAUSAL AGENTS

Most documented cases of human acanthocephaliasis are caused by *Macracanthorhynchus hirudinaceus*, *Macracanthorhynchus ingens*, and *Moniliformis moniliformis*. The true prevalence of these species in nature is not well understood, but *M. hirudinaceus* and *M. moniliformis*

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TABLE 1 Summary of cases reports of human acanthocephaliasis published in English language literature

Species ^b	Location	Symptom(s)	Treatment	Reference
<i>Acanthocephalan</i> , NOS (nr.)	United Kingdom	Eye discomfort	NA ^a	16
<i>Plagiorhynchus</i>				
<i>Bolbosoma</i> cf. <i>capitum</i>	Japan	Found incidentally on colonoscopy (colon cancer)	NA	3
<i>Bolbosoma nipponicum</i>	Japan	Found incidentally on routine enteroscopy (Crohn's disease)	NA	13
<i>Bolbosoma</i> sp.	Japan	Right lower quadrant pain mimicking appendicitis	NA	42
<i>Bolbosoma</i> sp.	Japan	Severe abdominal pain, nausea, ileus	NA	8
<i>Bolbosoma</i> sp.	Japan	Right lower quadrant pain mimicking appendicitis, abdominal perforation	NA	41
<i>Corynosoma</i> cf. <i>validum</i>	Japan	Abdominal pain, ileal ulcerations	Pyrantel pamoate	12
<i>Corynosoma villosus</i>	Japan	Abdominal pain, bloody stool (not loose)	NA	6
<i>Macracanthorhynchus hirudinaceus</i>	China	2 pediatric cases: acute abdominal pain, abdominal perforations, surgical removal	NA	23
<i>Macracanthorhynchus hirudinaceus</i>	China	Multicase review	NA	24
<i>Macracanthorhynchus hirudinaceus</i>	Thailand	Abdominal pain/tenderness, anorexia, nausea; progression to severe abdominal pain before admission and diarrhea	NA	19
<i>Macracanthorhynchus hirudinaceus</i>	Thailand	Abdominal pain, anorexia, nausea, intestinal perforation, ovarian cysts found during surgery	NA	20
<i>Macracanthorhynchus hirudinaceus</i>	Thailand	Abdominal pain, vomiting, fever, intestinal perforations	NA	21
<i>Macracanthorhynchus ingens</i>	USA (Florida)	Eosinophilia (8%)	Pyrantel pamoate (11 mg/kg)	10
<i>Macracanthorhynchus ingens</i> (as <i>M. hirudinaceus</i>)	USA (Louisiana)	NA	Pyrantel pamoate	7
<i>Macracanthorhynchus ingens</i>	USA (Ohio)	NA	Pyrantel pamoate (11 mg/kg)	5
<i>Macracanthorhynchus ingens</i>	USA (Texas)	NA	Pyrantel pamoate (11 mg/kg)	4
<i>Macracanthorhynchus ingens</i>	USA (Texas)	NA	Niclosamide + mebendazole	27
<i>Macracanthorhynchus</i> sp.	Papua New Guinea	Pediatric death associated with dysentery; worm found postmortem	NA	25
<i>Moniliformis moniliformis</i>	Australia	NA	Niclosamide	26
<i>Moniliformis moniliformis</i>	Iran	Diarrhea, vomiting, abdominal pain, coinfection with <i>Giardia duodenalis</i>	Levamisole	32
<i>Moniliformis moniliformis</i>	Iran	NA	Levamisole	9
<i>Moniliformis moniliformis</i>	Iran	Irritability, diarrhea, pallor, stunted growth, coinfection with <i>Giardia duodenalis</i>	Thiabendazole	33
<i>Moniliformis moniliformis</i>	Iran	Abdominal distention, anorexia, vomiting, diarrhea, stunted growth, eosinophilia (23%)	Niclosamide	34
<i>Moniliformis moniliformis</i>	Iran	Mild abdominal discomfort	Piperazine citrate	35
<i>Moniliformis moniliformis</i>	Iraq	Anorexia, diarrhea, weight loss, abdominal distention, coinfection with <i>Giardia</i>	NA	36
<i>Moniliformis moniliformis</i>	Nigeria	19 cases: diarrhea, abdominal discomfort	Ivermectin	14
<i>Moniliformis moniliformis</i>	Nigeria	Weakness, giddiness, burning sensation in umbilicus, loose stools, abdominal discomfort	Niclosamide	29
<i>Moniliformis moniliformis</i>	Zimbabwe (as Rhodesia)	Anorexia, abdominal discomfort, irritability	Mebendazole	30
<i>Moniliformis moniliformis</i>	Saudi Arabia	Anorexia	Mebendazole	31
<i>Moniliformis moniliformis</i>	Australia (Tasmania)	Cough, irritability	Paracetamol	37
<i>Moniliformis moniliformis</i>	USA (Florida)	NA	Pyrantel pamoate (11 mg/kg)	28
<i>Moniliformis moniliformis</i>	USA (Florida)	NA	Pyrantel pamoate (11 mg/kg)	11

^aNA, not available.^bNOS, not otherwise specified; nr., near.

probably occur nearly worldwide wherever pigs and rats occur, respectively. *Macracanthorhynchus ingens* is endemic to eastern and midwestern North America where raccoons occur (10). Other species that have been only very rarely implicated in human infection (with their natural distributions in parentheses) include *Acanthocephalus rauschi* (Alaska), *Pseudoacanthocephalus bufonis* (Southeast Asia), *Corynosoma* species (worldwide, marine), and

Bolbosoma species (worldwide, marine) (15). There is one report of an avian acanthocephalan (*Plagiorhynchus* or related) from the eye of a groundskeeper in England; however, it is believed that the presence of the parasite in the eye was incidental after accidental inoculation rather than true infection following consumption of an infected host (16).

BIOLOGY AND LIFE CYCLE

Acanthocephalans have a complex multihost life cycle, and only a few species have had their entire life cycles described. The three most common species implicated in human disease, *M. hirudinaceus*, *M. ingens*, and *M. moniliformis*, use pigs, raccoons, and rodents as their primary definitive hosts, respectively, although other carnivores may function as definitive hosts. Fully embryonated eggs containing an infective first-stage larva (acanthor) are shed in the feces of the definitive host. The eggs are ingested by an arthropod intermediate host. The primary groups of arthropods that serve as intermediate hosts include scarabaeoid beetles for *M. hirudinaceus*, millipedes for *M. ingens*, and various beetles and cockroaches for *M. moniliformis*; however, specificity for the arthropod host is believed to be low. Within the arthropod host, the parasite goes through multiple stages, from acanthor to acanthella to cystacanth, the last of which is the infective stage for the definitive host. The definitive host, including humans, becomes infected after ingesting arthropods containing infective cystacanths. In some cases, infection can occur from the ingestion of paratenic hosts. In the definitive host, liberated parasites attach to the mucosa of the small intestine, where they mature into adults and mate in about 8 to 12 weeks (17) (Fig. 1). The preponderance of human cases of *Macracanthorhynchus* and *Moniliformis* involving children may be due to children's habit of putting objects, including insects, in their mouths. Human infection has also been documented in communities that use insects and other arthropods for medicinal purposes (10).

EPIDEMIOLOGY OF HUMAN ACANTHOCEPHALIASIS

Acanthocephaliasis may be an ancient disease of humans. Analysis of fossilized feces from prehistoric humans in Utah have revealed the eggs of *Moniliformis*; however, it is unknown whether this represented true or spurious infections (18).

Macracanthorhynchus hirudinaceus is a nearly cosmopolitan parasite of pigs. Human infection has been documented in Thailand (19–22), Bulgaria (15), the Czech Republic (15), Vietnam (15), China (23, 24), Russia (15), Papua New Guinea (25), Australia (26), Madagascar (15), and Brazil (15). It has also been documented once from a patient in the United States (Louisiana) (7); however, two of us (M.R.C. and B.A.M.) had the opportunity to reexamine the specimen and determined it to be *M. ingens*. Human infection is most commonly seen in young children. Two cases were reported in children in China, ages 2.5 and 7 years old, presenting with acute abdominal colic (23). In a case from Thailand, *M. hirudinaceus* was recovered postmortem in a 32-year-old female who had died of rheumatic heart disease (22). In another case from Thailand, an adult worm was removed from the duodenum of a 26-year-old male patient by laparotomy (19). In the 1800s human infection was commonly reported in the Volga Valley in Russia, where the cockchafer beetle (*Melolontha melolontha*) was commonly consumed raw (23). The cases from Brazil were believed to be spurious following consumption of infected pig intestines (15).

Macracanthorhynchus ingens is a raccoon parasite endemic to eastern North America. To date there have been four documented human cases (exclusive of the aforementioned case from Louisiana) from Texas (4, 27), Florida (10), and Ohio (5). The reported cases in the United States have all been in children under the age of 18 months and diagnosed by the finding of adult worms in stool. The two cases in Texas were reported for infants of the ages 10 months and 15 months (4, 27). The case from Florida was reported for an 18-month-old female. The patient was asymptomatic but presented with eosinophilia. There were no known animal contacts, but the parents reported millipedes, the natural intermediate host for *M. ingens*, around the yard and garden (10). The infection in Ohio occurred in a 17-month-old female who was examined by her primary care physician after a parent noticed the worm in her stool. This Ohio case was novel in that eggs of *M. ingens* were also detected



Acanthocephala

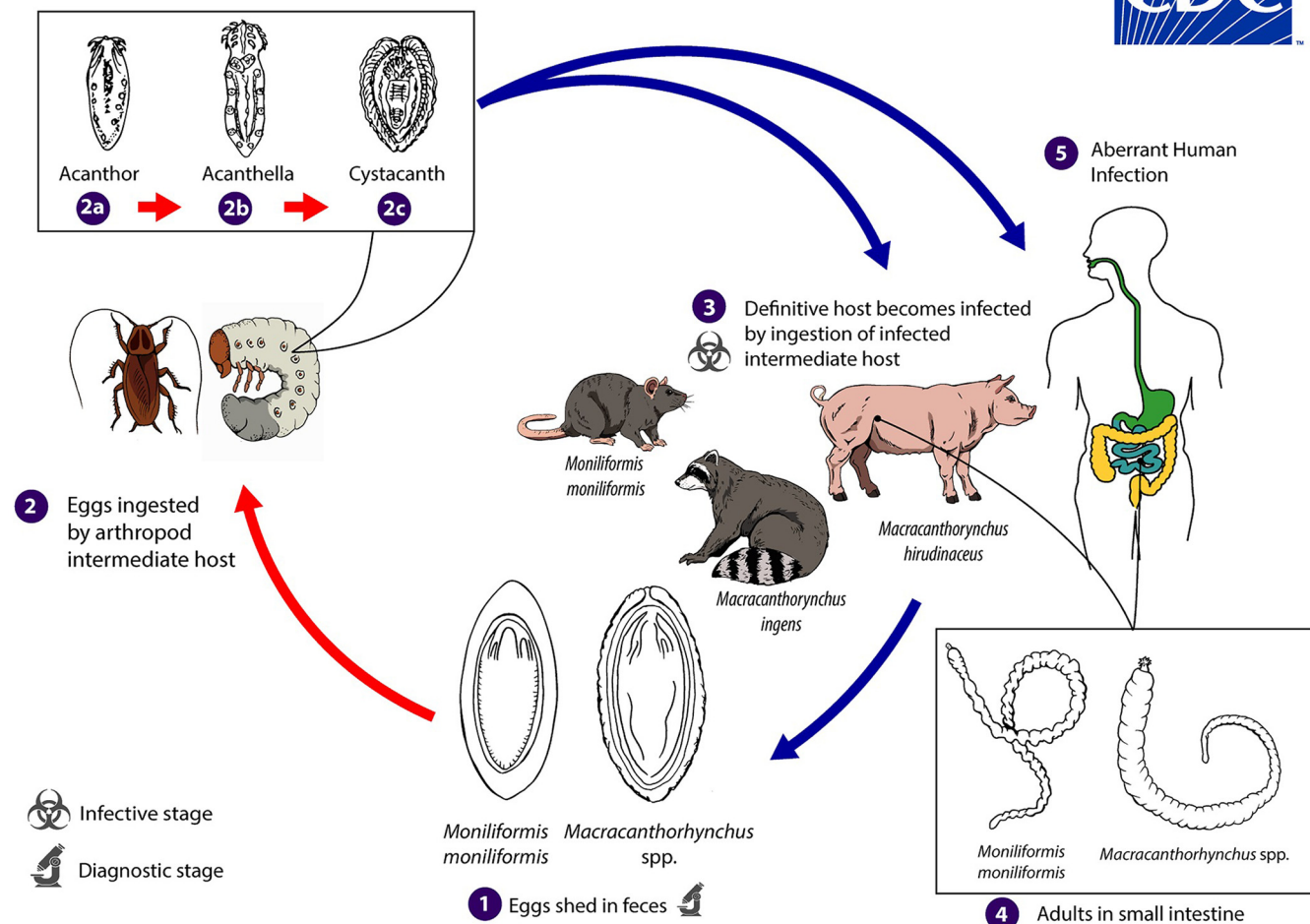


FIG 1 Life cycle of acanthocephalans. Eggs are shed in the feces of the definitive hosts (1). The eggs contain a fully developed acanthor when shed in feces. The eggs are ingested by an intermediate host (2), which is an arthropod. Within the hemocoel of the insect, the acanthor (2a) molts into a second larval stage, called an acanthella (2b). After 6 to 12 weeks, the worm reaches the infective stage, called a cystacanth (2c). The definitive host becomes infected upon ingestion of intermediate hosts containing infective cystacanths (3). In the definitive host, liberated juveniles attach to the wall of the small intestine, where they mature and mate in about 8 to 12 weeks. In humans (4), the worms seldom develop to full maturity and produce eggs. (Courtesy of the CDC-DPDx.)

by ova-and-parasite (O&P) examination, indicating patent infection. The patient had no history of gastrointestinal symptoms or changes in bowel habit. She lived in rural Ohio and had no travel history outside the region (5). In the case from Louisiana, the patient, a 17-month-old male, had no documented animal exposures but regularly played outside, where he was reportedly seen putting objects in his mouth (7).

Moniliformis moniliformis is a cosmopolitan parasite of rats and other rodents. Human infection has been reported from the United States (11, 28), Belize (15), Colombia (15), Italy (15), Egypt (15), Nigeria (14, 29), Sudan (15), Zimbabwe (as Rhodesia) (30), Zambia (15), Saudi Arabia (31), Iran (9, 32–35), Iraq (36), Israel (15), Russia (15), Bangladesh (15), and Australia (26). Most cases are in children at or under 2 years of age (5, 10). *Moniliformis moniliformis* has been reported from Iran at least seven times, and in two cases, the patients were coinfecting with *Giardia duodenalis* (35, 36). In one of those two, the patient was diagnosed with pica and the mother indicated that the child had been observed putting cockroaches in her mouth (35). A case diagnosed from Tasmania in a 14-month-old male was believed to have been acquired on mainland Australia (37). In 2006, a 20-month-old female from the Eastern Province of Saudi Arabia was diagnosed with *M. moniliformis*. The parents reported that one

or two worms were seen daily in the patient's stool for the prior 2 months; the mother also noted the patient had been observed on several occasions ingesting cockroaches (31).

Human infection with *Acanthocephalus rauschi* has been reported in French language literature (38) but could not be reviewed by us. *Acanthocephalus rauschi* was described from the Alaskan grayling (*Thymallus arcticus*) and North American river otter from Alaska (39), but the source of human infection is unknown.

Pseudoacanthocephalus bufonis, a parasite of toads in Southeast Asia, has been implicated in human infection in Indonesia. The intermediate or paratenic hosts, which are the likely source of human infection, are unknown, and the single human case was diagnosed postmortem on autopsy (40).

Members of the genus *Bolbosoma* are parasites of whales. Marine planktonic crustaceans are believed to serve as the intermediate host and fish as paratenic hosts. Human infection is believed to be acquired from the consumption of infected fish. All human cases of *Bolbosoma* infection to date have been documented from Japan (3, 8, 13, 41, 42). In one case, the patient had reported eating sashimi as part of his diet; in that case the parasite was identified as *Bolbosoma* cf. *capitatum* by DNA sequencing (3). In another case, a worm was removed from the jejunum of a 27-year-old female with bowel obstruction and identified by DNA sequencing as *Bolbosoma* (8). In yet another case, *Bolbosoma* was recovered from a 37-year-old male with a history of Crohn's disease; the worm was removed surgically from the jejunal mucosa and was identified by DNA sequencing as *Bolbosoma nipponicum* (13). A 16-year-old boy was diagnosed with a probable *Bolbosoma* species infection after the recovery of an acanthocephalan in a tumor in the serosa over the ileum (42).

Members of the genus *Corynosoma* use marine amphipod crustaceans as intermediate hosts and pinnipeds and whales as definitive hosts, with marine fish serving as paratenic hosts and the source of infection. Two human cases have been recorded from Japan (6, 12) and one has been recorded from Alaska (43). The case from Alaska was diagnosed by microscopy of the intact worm from an Inuit male; the parasite was not identified to the species level (43). In one case from Japan, a worm was removed from the ileum of a 73-year-old male with a 2-year history of abdominal pain and a history of consuming uncooked seafood; the parasite was identified as *Corynosoma villosum* by molecular methods (6). In the other case from Japan, a worm was removed from the ileum of a 70-year-old female and was identified by DNA sequencing as *Corynosoma* cf. *validum* (12).

CLINICAL PRESENTATION, PATHOLOGY, AND TREATMENT

The majority of diagnosed cases of acanthocephaliasis are associated with no overt symptomatology (3–5, 7, 9–11, 13, 26–28). Passage of an adult worm is typically the trigger for clinical evaluation, as the worms are large and readily noticeable in a stool specimen. Cases have also been diagnosed incidentally during endoscopic examination for nondescript gastrointestinal symptoms or other preexisting medical conditions, such as colon cancer, Crohn's disease, and ovarian cysts (3, 13, 20). The overall spectrum of symptoms associated with acanthocephaliasis can be quite wide, and peripheral eosinophilia may be present in only some cases despite eosinophilic infiltration in tissues (10, 34). Few published cases of asymptomatic shedding of adult worms include peripheral white blood cell count, and those that do rarely include eosinophil count in the differential. Mild symptoms can include anorexia, nausea, cramping, constipation, diarrhea, abdominal discomfort, and irritability in young children (14, 29–31, 33, 35, 37). More moderate symptoms can include vomiting, severe abdominal pain, abdominal distension, and weight loss (12, 19, 32, 34, 36, 42). Severe symptoms include bloody diarrhea and ileus, perforation, and/or ulceration of the intestinal wall (most common with *Bolbosoma* and *Corynosoma*) (6, 8, 20, 21, 23, 41). Rare extraintestinal manifestations have also been reported (described above) (16).

The pathology within the intestinal tract has been described in multiple cases to date, each with unique aspects depending on the clinical severity (8, 19, 21). The typical findings at the site of attachment of the proboscis are characterized by ulceration of the

mucosal surface with profound white blood cell infiltration which may include neutrophils, eosinophils, and/or lymphocytes (15, 44). Lymphocytic and monocytic infiltration has also been described at sites of submucosal edema (8, 19). Lacteals in the lamina propria are dilated with hemorrhage in the ileal wall, which may resemble necrotizing enteritis (15, 44). Follicular hyperplasia may be seen in mesenteric lymph nodes, with eosinophils entering the lymph nodes resulting in dilatation.

Treatment for acanthocephalans is largely unstandardized. For mild or asymptomatic cases, many patients have been treated successfully with pyrantel pamoate (11 mg/kg of body weight per dose) orally in a three-dose regimen separated by 2 weeks per dose (4, 5, 10, 11, 28). Typically, patients ceased to shed worms, ceased to have symptoms, or were negative on fecal exam for eggs after treatment with pyrantel pamoate. Previous reports have also described treatment success with levamisole (9, 32), niclosamide (26, 29, 34), mebendazole (30, 31), ivermectin (14), paracetamol (37), thiabendazole (33), and piperazine citrate (35). For more invasive infections involving bowel perforations, ileus, or ulceration, endoscopic removal of the worm without concomitant antihelminth therapy is common (8, 20, 21, 23, 41, 42).

DIAGNOSIS AND IDENTIFICATION

Acanthocephaliasis is usually diagnosed by the identification of adult parasites in stool specimens, usually in diaper-aged children. The number of cases with this particular demographic is probably largely due to (i) the habits of children to eat insects and (ii) the fact that a diaper may serve as a good collection apparatus, especially with astute parents who are likely to observe the worms when changing diapers. In children who are toilet trained, any evidence may be inconveniently flushed away. Patent acanthocephalan infections in humans are rare but do occur, and in those instances identification can be made by the finding of eggs in concentrated wet mounts of stool (5). Many technologists may not be trained to immediately identify the eggs; however, the eggs have a distinctive morphology that should draw attention and warrant further scrutiny.

Adult acanthocephalans are large, pseudocoelomate worms. Female specimens of *M. moniliformis* measure 10 to 27 cm; males are smaller, at 4 to 10 cm (Fig. 2A). Female specimens of *Macracanthorhynchus* measure 12 to 32 cm; males are smaller, at 7 to 8 cm (Fig. 2B). Adults of both genera are long and slender. They superficially resemble adults of *Ascaris lumbricoides* but are usually heavily wrinkled (*Macracanthorhynchus*) or exhibit pseudosegmentation (*Moniliformis*), and they do not exhibit a consistent length-to-width ratio along the length of the body. The anterior ends of both genera have readily visible proboscis receptacles which are hollow round openings, compared to the three "lips" seen with ascarids. The armed proboscis may be extended or retracted, and when retracted, some dissection may be necessary for confirmation and identification (10). Separation of *M. hirudinaceus* from *M. ingens* requires morphometric analysis of the proboscis and hooklets (45) or molecular methods such as PCR followed by sequencing analysis of the cytochrome *c* oxidase subunit I (COX-I) gene (5).

Eggs of *M. moniliformis* measure 90 to 125 μm long by 65 μm wide and have a thin shell; the mature acanthor is visible within the egg (Fig. 2C). Eggs of *Macracanthorhynchus* measure 80 to 100 μm long by 50 μm wide and possess a thick, wrinkly, and deeply pitted shell; internally a mature acanthor is visible (Fig. 2D). The eggs of *M. hirudinaceus* and *M. ingens* cannot be separated morphologically (10).

Acanthocephalans can also be identified by histologic examination (Fig. 2E). The tegument is thick and both circular and longitudinal muscles are present. There is no digestive tract, and the reproductive tract occupies most of the body cavity. The reproductive organs are contained within a connective tissue sheath called a ligament sac. Female worms may have one or two ovaries that are typically broken into sections called ovarian balls; males typically have two testes (44). The proboscis, when embedded in host tissue, usually displays hooklets (Fig. 2E).

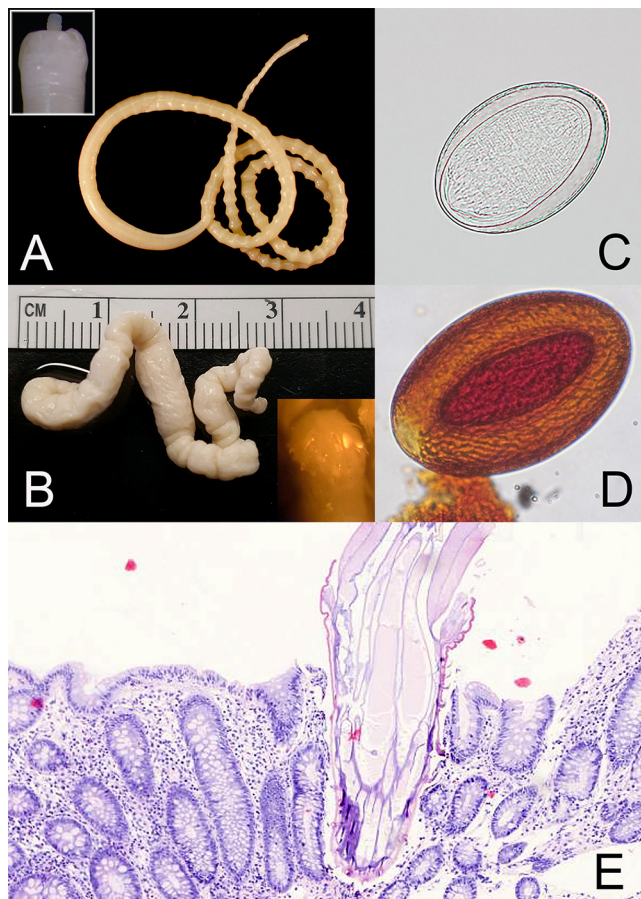


FIG 2 Adults and eggs of acanthocephalans. (A) *Moniliformis moniliformis*, adult; inset shows closeup of the extended proboscis. (B) *Macracanthorhynchus ingens*, adult; inset shows closeup of retracted proboscis after dissection. (C) Unstained wet mount of an egg of *Moniliformis moniliformis*. (Courtesy of the CDC-DPDx.) (D) Iodine-stained wet mount of an egg of a *Macracanthorhynchus* sp. (E) Proboscis of an acanthocephalan embedded in human intestinal epithelium. (Courtesy of Ramon Sandin.)

Human infection and clinical symptomology due to acanthocephalans are rarely reported. This likely poses a problem in the appropriate detection and identification of these organisms. Due to the lack of familiarity of acanthocephalans among trained microscopists, expert consultation may be needed for these cases. Laboratorians may want to consider consulting their local and state public health laboratories, large reference laboratories with expert parasitologists on staff, or the Centers for Disease Control and Prevention, which offers a specialized service for parasite diagnostics.

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